NORMAL HEME SYNTHESIS

ALA synthase Δ AMINOLEVULINIC ACID ALA dehydratase PORPHOBILINOGEN porphobilinogen dehydratase **HYDROXYMETHYLBILANE** uroporphyrinogen synthase + cosynthase spontaneous UROPORPHYRINOGEN I UROPORPHYRINOGEN III uroporphyrinogen decarboxylase COPROPORPHYRINOGEN III COPROPORPHYRINOGEN I coproporphyrinogen oxidase **PROTOPORPHYRINOGEN** protoporphyrinogen oxidase PROTOPORPHYRIN ferrochelatase and iron

HEME

Uroporphyrin I (URO I)

UROPORPHYRIN III (URO III)

△ AMINOLEVULINIC ACID (ALA)

TOOC—
$$CH_2-CH_2-C-S-CoA + TOOC-CH_2-NH_3^+$$
succinyl-CoA

Aminolevulinic
Acid Synthase

CoA-SH

CoA-SH

CoA-SH

CoA-SH

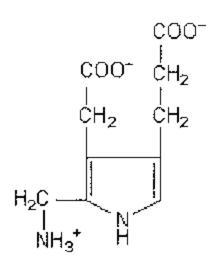
CoA-SH

CoA-SH

CoA-SH

Aminolevulinate (ALA)

PORPHOBILINOGEN (PBG)



HBMS HEME SYNTHESIS

ALA synthase A AMINOLEVULINIC ACID ALA dehydratase PORPHOBILINOGEN porphobilinogen dehydratase HYDROXYMETHYLBILANE overproduction of urogen III synthase + cosynthase spontaneous **UROGEN III** UROGEN I overproduction of urogen decarboxylase COPROGEN I COPROGEN III coproporphyrinogen oxidase PROTOPORPHYRINOGEN protoporphyrinogen oxydase **PROTOPORPHYRIN** ferrochelatase and iron HEME